Pediatrics and Neonatology (2016) 57, 232–235

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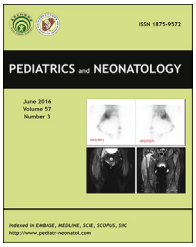

CASE REPORT

Neonatal Sigmoid Colon Perforation: A Rare Occurrence in Low Anorectal Malformation and Review of the Literature

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Received Sep 22, 2012; received in revised form Jul 1, 2013; accepted Jul 22, 2013
Available online 11 November 2013

Key Words
anorectal malformations;
perforative peritonitis;
pneumoperitoneum;
sigmoid colon

Gastrointestinal perforation in neonates with anorectal malformation (ARM) is extremely uncommon. Delayed patient presentation is an important causative factor. A 2.5-kg neonate presented 72 hours after birth with abdominal distention and absent anal opening with meconium pearls. An abdominal X-ray revealed the presence of free gas. After adequate resuscitation patient underwent surgery. Closure of the sigmoid colon perforation with a proximal diverting loop colostomy with anoplasty was done. The literature reveals only two cases of sigmoid colon perforation with low ARM. Ours is the third case, in whom repair of the perforation and correction of the ARM was managed successfully at the same time.

1. Introduction

In infancy and childhood, perforation of the colon is second only to the ileum, but occurrence in the neonatal period is rare.[1] Physical examination of the perineum is often sufficient to diagnose anorectal malformation (ARM) in neonates. However, delay in diagnosis is not uncommon, even in developed countries, diagnostic delays of 3–43 days have been reported in as many as 21–32% of newborns.[2] In developing countries, initiation of treatment is further delayed by social factors such as poverty, illiteracy, poor transport facilities, and scarcity of specialists.[3] Hirschsprung's disease, enterocolitis, and instrumentation are commonly described etiological factors but anorectal malformations are very rare.[1] A high index of suspicion in neonates with ARM presenting with sepsis and features of peritonitis such as a tense distended abdomen with parietal wall edema and erythema may lead to diagnosis.[4] The type of surgical intervention depends upon the physiological

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http://dx.doi.org/10.1016/j.pedneo.2013.07.011

state of patient, site of perforation, type of anorectal anomaly, and degree of peritoneal contamination.[4]

2. Case Report

A 2.5-kg male child presented 72 hours after birth with increasing abdominal distension and absent anal opening. The baby cried soon after birth and passed clear urine within 24 hours. However, he had not passed meconium for 3 days, for which he was referred to us on the 4th day after birth. On physical examination, the child was found to be dyspneic, lethargic, and dehydrated. The abdomen was distended and tender. There was no erythema or edema of the anterior abdominal wall. Perineal examination revealed the presence of meconium pearls and absent anal opening, suggestive of low ARM, as shown in Figure 1. The penis was normal with the urethral opening at the tip, and both testes were palpable in the scrotum. Meconium pearls were extending along the median raphe up to the penoscrotal junction. An abdominal X-ray suggested a large saddle-shaped air shadow below the diaphragm indicating pneumoperitoneum, as shown in Figure 2. Hematological investigations were within normal limits. The child was resuscitated with intravenous fluids and antibiotics were started.

After stabilizing his general condition, an exploratory laparotomy was done using a supra-umbilical right transverse incision. A gush of air with meconium was noticed with fibrinous flakes over the loops of small bowel and colon. A longitudinal perforation of 2 × 1 cm was noticed in the lower sigmoid colon with meconium coming out, as shown in Figure 3. There was no evidence to suggest the concurrent presence of necrotizing enterocolitis. The perforation was closed in two layers. A thorough peritoneal lavage was given and a proximal diverting loop colostomy was done in view of gross contamination. Anoplasty for low ARM was also done. The postoperative period was

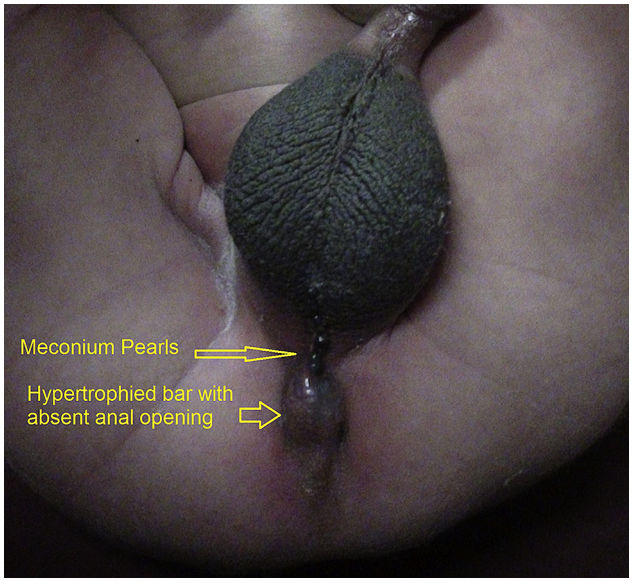

Figure 1 Preoperative photograph showing meconium pearls suggestive of low anorectal malformation.

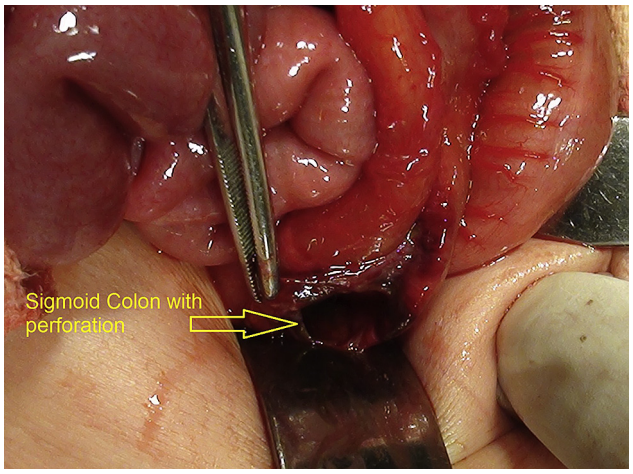

Figure 2 Intra-operative photograph showing sigmoid colon perforation.

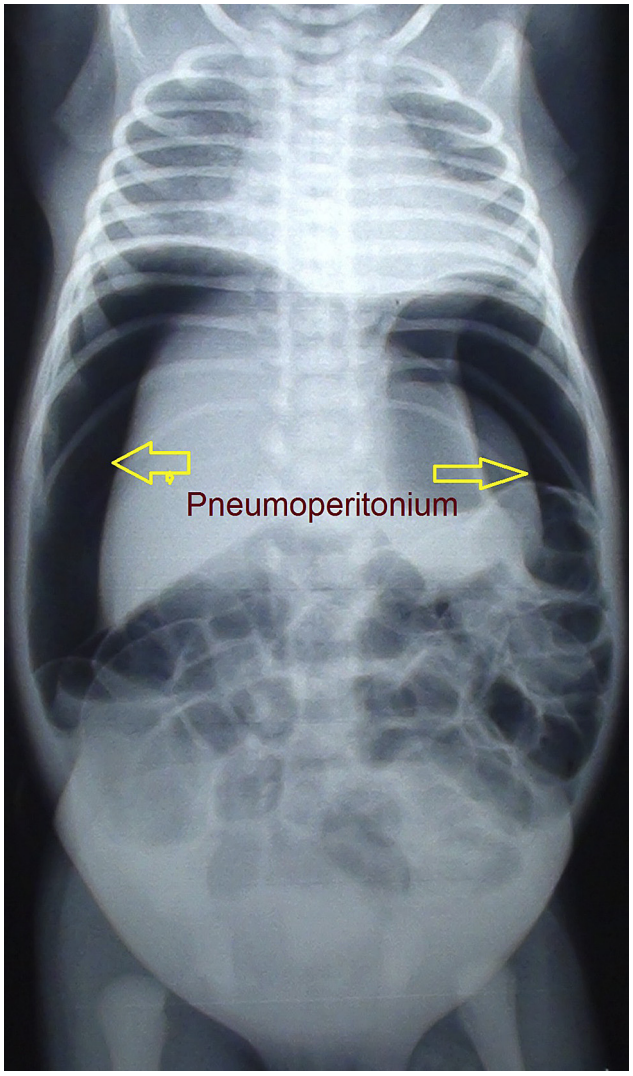

Figure 3 Plain X-ray abdomen showing free gas under both domes of diaphragm suggestive of perforative peritonitis.

Table 1 Summary of literature on sigmoid colon perforation complicating low anorectal malformation (ARM).

Sr.No.	Author (year)	Sex	Age (h)	ARM type	Site of perforation	Peritoneal soiling	Management of perforation	Outcome
1	Amundsen (1958)	Male	36	Low—no fistula	Sigmoid colon	Contained	Anoplasty + PSEC	Survived
2	Sharma (2004)	Male	144	Low—no fistula	Sigmoid colon	—	PSEC	Survived
3	Sandesh Parelkar (2012)	Male	72	Low—no fistula	Sigmoid colon	Diffuse	Anoplasty CP + PC	Survived

Age = age at diagnosis of pneumoperitoneum or onset of perforation; CP + PC = closure of perforation + proximal colostomy; PSEC = perforation site exteriorized as colostomy.

uneventful. The patient is on regular anal dilatation and has satisfactory weight gain.

3. Discussion

Spontaneous perforation of the colon is estimated to occur in 2% of neonates with ARM and the incidence rises to 9.5% when the diagnosis is delayed.[2] Colonic perforations account for 15% of pneumoperitoneum seen in the neonatal age group.[5] Bowel perforation increases neonatal mortality of ARM from 3% to 23%.[5] The literature on this topic is restricted to anecdotal information and isolated case reports.[6–10] Perforation of the colon in the newborn is a serious and rare complication. Perforations secondary to anorectal malformations are very rare as these malformations are generally diagnosed early and treated before the perforation can occur.

A review of the literature revealed approximately 63 cases of ARM with perforation. However, adequate details are available in only 26 cases. Only two cases of sigmoid colon perforation with low ARM with no fistula have been reported previously; ours is the third case. The relative paucity of the literature on spontaneous perforation of the colon in ARM is due to the rarity of its occurrence and inadequate reporting. The exact incidence of bowel perforation in ARM is not known. The median age at the onset or diagnosis of perforation in ARM cases was 48 hours.[2,9] Turowski et al[2] reported two perforations among 99 cases of ARM (2% incidence). In a subset analysis, they found two perforations (9.5%) among 21 cases with delayed presentation of ARM. Mathur et al[11] recorded five perforations (6.5%) among 77 cases of ARM with congenital pouch colon (CPC). An Indian study[12] reported two perforations (1.6%) of 125 ARM. High and low varieties of ARM are equally susceptible to perforation. Bowel rupture frequently occurs in ARM without fistula; however, anomalies with fistula are not spared. Occlusion of a tiny fistula by inspissated meconium may have caused perforation due to raised intraluminal pressure in cases of fistulous ARM (usually decompressed by fistula).[11] Nearly 85% of perforations occurred in boys and the rarity of perforation in females is probably caused by the high frequency of low ARM with a wide rectoforchette fistula.

The etiopathogenesis of gastrointestinal perforation neonates with ARM may be explained by a combination of factors.[4] The downstream occlusion results in proximal intestinal dilatation and increase in intraluminal pressure resulting in tension gangrene. It may undergo perforation even when the closed loop obstruction has been relieved, precipitating an ischemia—reperfusion injury which should emphasize the vital role of close clinical observation of such cases in the postoperative period.[4] The cecum is the most common site. A high index of suspicion in neonates with ARM presenting with sepsis and features of peritonitis should be noted. Although features of pneumoperitoneum on abdominal X-ray have been reported in 60—70% of neonates with gastrointestinal perforation, its presence is confirmatory.[4] The management of gastrointestinal perforation in neonates with ARM aims at aggressive resuscitation and early surgical intervention.[4] Primary closure of the perforation may be attempted in selective cases; exteriorization of the perforation as a stoma or its primary closure with a proximal diverting stoma can be an option.

Turowski et al provided additional evidence that delayed diagnosis of anorectal malformation (ARM) appears to be a common problem, occurring in 21% of their patients. Delayed diagnosis of anorectal malformations has become increasingly familiar to surgeons, as evidenced by the number of publications on this topic in the literature.[2] Clearly, the cornerstone to timely diagnosis of an ARM continues to be a comprehensive neonatal examination performed by a pediatrician or a pediatric trainee with sufficient experience.

The overall mortality of perforated ARM is 19%. Sepsis and disseminated intravascular coagulation were frequent causes of death. Better understanding of the pathophysiology of perforation in ARM and early surgical decompression of the obstructed colon can be expected to reduce mortality in future.[13] Only two cases of sigmoid colon perforation presenting as a complication of low ARM have been reported.[6,14] The differences in those two cases were found at the time of presentations and management [Table 1: Sharma — 144 hours; PSEC (perforation site exteriorized as colostomy), Amundsen — 36 hours; anoplasty + PSEC, our case — 72 hours; anoplasty + PSEC].

Diagnosis of ARM by comprehensive neonatal examination performed by a pediatrician or a pediatric trainee and early surgical decompression can reduce mortality. Primary closure of perforation with proximal colostomy is a safe treatment option in presence of gross contamination as in our patient. Correction of low ARM with perforation can also be undertaken at the same time.

References

1. Rikham PP, Lister J, Irving IM, editors. *Necrotizing enterocolitis: bacterial and meconium peritonitis. Neonatal surgery.*

2nd ed. London: Butterworth Public Company; 1978. p. 411—28.
2. Turowski C, Dingemann J, Gillick J. Delayed diagnosis of imperforate anus: an unacceptable morbidity. *Pediatr Surg Int* 2010;**26**:1083—6.
3. Adejuyigbe O, Abubakar AM, Sowande OA, Olayinka OS, Uba AF. Experience with anorectal malformations in Ile-Ife, Nigeria. *Pediatr Surg Int* 2004;**20**:855—8.
4. Digray NC, Mengi Y, Goswamy HL, Thappa DR. Colorectal perforations in neonates with anorectal malformations. *Pediatr Surg Int* 2001;**17**:42—4.
5. Khan TR, Rawat JD, Ahmed I, Rashid KA, Maletha M, Wakhlu A, et al. Neonatal pneumoperitoneum: a critical appraisal of its causes and subsequent management from a developing country. *Pediatr Surg Int* 2009;**25**:1093—7.
6. Sharma SB, Gupta V, Sharma V. Gastrointestinal perforations in neonates with anorectal malformations. *Indian J Gastroenterol* 2004;**23**:107—8.
7. Komuro H, Urita Y, Hori T, Hirai M, Kudou S, Gotoh C, et al. Perforation of the colon in neonates. *J Pediatr Surg* 2005;**40**: 1916—9.
8. Fares AE, El-Saket HM. Neonatal colonic perforation: a case series. *Arab J Gastroenterol* 2008;**9**:75—9.
9. Eltayeb AA. Delayed presentation of anorectal malformations: the possible associated morbidity and mortality. *Pediatr Surg Int* 2010;**26**:801—6.
10. Sandlas G, Kothari P, Sarda D, Karkera P. Spontaneous bowel perforation in a neonate with anorectal malformation. *Saudi J Gastroenterol* 2011;**17**:72—3.
11. Mathur P, Saxena AK, Bajaj M, Chandra T, Sharma NC, Simlot A, et al. Role of plain abdominal radiographs in predicting type of congenital pouch colon. *Pediatr Radiol* 2010; **40**:1603—8.
12. Chalapathi G, Chowdhary SK, Rao KL, Samujh R, Narasimhan KL, Mahajan JK, et al. Risk factors in the primary management of anorectal malformations in Northern India. *Pediatr Surg Int* 2004;**20**:408—11.
13. Raveenthiran V. Spontaneous perforation of the colon and rectum complicating anorectal malformations in neonates. *J Pediatr Surg* 2012;**47**:720—6.
14. Amundsen P. Rupture of the sigmoid colon in a newborn infant with imperforate anus. *Br J Radiol* 1958;**31**:47—9.